F13B gene

coagulation factor XIII B chain

Normal Function

The *F13B* gene provides instructions for making one part, the B subunit, of a protein called factor XIII. This protein is part of a group of related proteins called coagulation factors that are essential for normal blood clotting. They work together as part of the coagulation cascade, which is a series of chemical reactions that forms blood clots in response to injury. After an injury, clots seal off blood vessels to stop bleeding and trigger blood vessel repair. Factor XIII acts at the end of the cascade to strengthen and stabilize newly formed clots, preventing further blood loss.

Factor XIII in the bloodstream is made of two A subunits (produced from the *F13A1* gene) and two B subunits (produced from the *F13B* gene). The role of the B subunits is to carry and stabilize the A subunits, protecting them from being broken down. When a new blood clot forms, the A and B subunits separate from one another, and the A subunits are cut (cleaved) to produce the active form of factor XIII (factor XIIIa). The active protein links together molecules of fibrin, the material that forms the clot, which strengthens the clot and keeps other molecules from breaking it down.

Studies suggest that factor XIII has additional functions, although these are less well understood than its role in blood clotting. Specifically, factor XIII is likely involved in other aspects of wound healing, immune system function, maintaining pregnancy, bone formation, and the growth of new blood vessels (angiogenesis).

Health Conditions Related to Genetic Changes

age-related macular degeneration

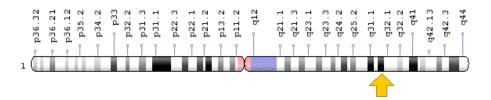
factor XIII deficiency

At least 17 mutations in the *F13B* gene have been found to cause inherited factor XIII deficiency, a rare bleeding disorder. Without treatment, affected individuals have a greatly increased risk of abnormal bleeding episodes, including life-threatening bleeding inside the skull (intracranial hemorrhage). *F13B* gene mutations severely reduce the amount or disrupt the function of the B subunit of factor XIII, preventing it from stabilizing and protecting the A subunit. The resulting loss of factor XIII activity weakens new blood clots and prevents them from stopping blood loss effectively.

Chromosomal Location

Cytogenetic Location: 1q31.3, which is the long (q) arm of chromosome 1 at position 31.3

Molecular Location: base pairs 197,038,741 to 197,067,267 on chromosome 1 (Homo sapiens Annotation Release 108, GRCh38.p7) (NCBI)



Credit: Genome Decoration Page/NCBI

Other Names for This Gene

- coagulation factor XIII B chain precursor
- coagulation factor XIII, B polypeptide
- fibrin-stabilizing factor B subunit
- FXIIIB
- protein-glutamine gamma-glutamyltransferase B chain
- TGase
- transglutaminase B chain

Additional Information & Resources

Educational Resources

 Biochemistry (fifth edition, 2002): Diagram: Blood Clotting Cascade https://www.ncbi.nlm.nih.gov/books/NBK22589/?rendertype=figure&id=A1401

Scientific Articles on PubMed

PubMed

https://www.ncbi.nlm.nih.gov/pubmed?term=%28F13B%5BTIAB%5D%29+OR+%28FXIIIB%5BTIAB%5D%29+OR+%28%28coagulation+factor+XIII%5BTIAB%5D%29+AND+%28beta%5BTIAB%5D%29%29+OR+%28%28coagulation+factor+XIII%5BTIAB%5D%29+AND+%28B%5BTIAB%5D%29%29+AND+english%5Bla%5D+AND+human%5Bmh%5D+AND+%22last+1800+days%22%5Bdp%5D

OMIM

 FACTOR XIII, B SUBUNIT http://omim.org/entry/134580

Research Resources

- ClinVar https://www.ncbi.nlm.nih.gov/clinvar?term=F13B%5Bgene%5D
- Factor XIII Registry Database: F13B Gene http://www.f13-database.de/content.aspx?menu=1,5,10
- Factor XIII Registry Database: F13B Mutations http://www.f13-database.de/content.aspx?menu=1,6,24
- HGNC Gene Family: Sushi domain containing http://www.genenames.org/cgi-bin/genefamilies/set/1179
- HGNC Gene Symbol Report http://www.genenames.org/cgi-bin/gene_symbol_report?q=data/ hgnc_data.php&hgnc_id=3534
- NCBI Gene https://www.ncbi.nlm.nih.gov/gene/2165
- UniProt http://www.uniprot.org/uniprot/P05160

Sources for This Summary

- Bagoly Z, Koncz Z, Hársfalvi J, Muszbek L. Factor XIII, clot structure, thrombosis. Thromb Res. 2012 Mar;129(3):382-7. doi: 10.1016/j.thromres.2011.11.040. Epub 2011 Dec 24. Review. Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/22197181
- Biswas A, Ivaskevicius V, Seitz R, Thomas A, Oldenburg J. An update of the mutation profile of Factor 13 A and B genes. Blood Rev. 2011 Sep;25(5):193-204. doi: 10.1016/j.blre.2011.03.001. Epub 2011 Jun 2. Review.
 Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/21640452
- Biswas A, Ivaskevicius V, Thomas A, Oldenburg J. Coagulation factor XIII deficiency. Diagnosis, prevalence and management of inherited and acquired forms. Hamostaseologie. 2014;34(2):160-6. doi: 10.5482/HAMO-13-08-0046. Epub 2014 Feb 7. Review.
 Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/24503678
- OMIM: FACTOR XIII, B SUBUNIT http://omim.org/entry/134580
- Hethershaw EL, Cilia La Corte AL, Duval C, Ali M, Grant PJ, Ariëns RA, Philippou H. The effect of blood coagulation factor XIII on fibrin clot structure and fibrinolysis. J Thromb Haemost. 2014 Feb; 12(2):197-205. doi: 10.1111/jth.12455.
 - Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/24261582

- Levy JH, Greenberg C. Biology of Factor XIII and clinical manifestations of Factor XIII deficiency. Transfusion. 2013 May;53(5):1120-31. doi: 10.1111/j.1537-2995.2012.03865.x. Epub 2012 Aug 28. Review.
 - Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/22928875
- Muszbek L, Bereczky Z, Bagoly Z, Komáromi I, Katona É. Factor XIII: a coagulation factor with multiple plasmatic and cellular functions. Physiol Rev. 2011 Jul;91(3):931-72. doi: 10.1152/ physrev.00016.2010. Review.
 - Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/21742792
- Schroeder V, Kohler HP. Factor XIII deficiency: an update. Semin Thromb Hemost. 2013 Sep;39(6): 632-41. doi: 10.1055/s-0033-1353392. Epub 2013 Aug 8. Review.
 Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/23929307

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